

## EAST Search History

Ref #	Hits	Search Query	DBs	Default Operator	Plurals	Time Stamp
L1	1	Guy-John.IN.	US-PGPUB; USPAT; USOCR; EPO; JPO; DERWENT; IBM_TDB	OR	OFF	2006/12/01 09:57
L2	2	"ND4 mitochondrial protein"	US-PGPUB; USPAT; USOCR; EPO; JPO; DERWENT; IBM_TDB	OR	OFF	2006/12/01 09:57
L3	6	ND4 WITH mitochondria	US-PGPUB; USPAT; USOCR; EPO; JPO; DERWENT; IBM_TDB	OR	OFF	2006/12/01 09:58
L4	35	ND4 WITH (mitochondrial protein)	US-PGPUB; USPAT; USOCR; EPO; JPO; DERWENT; IBM_TDB	OR	OFF	2006/12/01 10:00
L5	67456	(mitochondrial codon) and (nuclear codon)	US-PGPUB; USPAT; USOCR; EPO; JPO; DERWENT; IBM_TDB	OR	OFF	2006/12/01 10:01
L6	79	L5 and ND4	US-PGPUB; USPAT; USOCR; EPO; JPO; DERWENT; IBM_TDB	OR	OFF	2006/12/01 10:08
L7	112	"Leber Hereditary Optic Neuropathy" and LHON	US-PGPUB; USPAT; USOCR; EPO; JPO; DERWENT; IBM_TDB	OR	OFF	2006/12/01 10:09
L8	16	L6 and L7	US-PGPUB; USPAT; USOCR; EPO; JPO; DERWENT; IBM_TDB	OR	OFF	2006/12/01 10:10

## EAST Search History

L9	7	G3460A or G11778A or T14484C	US-PGPUB; USPAT; USOCR; EPO; JPO; DERWENT; IBM_TDB	OR	OFF	2006/12/01 10:12
L10	5	L7 and L9	US-PGPUB; USPAT; USOCR; EPO; JPO; DERWENT; IBM_TDB	OR	OFF	2006/12/01 10:13

Kodaira, Tokyo 187-8502, Japan. goto@ncnp.go.jp  
SO Mitochondrion, (2003) Vol. 2, No. 4, pp. 293-304. .  
Refs: 34  
ISSN: 1567-7249 CODEN: MITOCN  
PUI S 1567-7249(03)00003-5  
CY Netherlands  
DT Journal; Article  
FS 005 General Pathology and Pathological Anatomy  
007 Pediatrics and Pediatric Surgery  
022 Human Genetics  
LA English  
SL English  
ED Entered STN: 3 Apr 2003  
Last Updated on STN: 3 Apr 2003  
AB A novel mitochondrial DNA point mutation, a C-to-A mutation at nucleotide position (np) 11,777, was identified in two unrelated patients out of 100 with Leigh syndrome. This mutation converted a highly evolutionary conserved arginine to a serine at codon 340 in ND4 gene. This codon was also converted by a G-to-A mutation at np 11,778, the most common mutation associated with Leber's hereditary optic neuropathy (LHON), but the amino acid replacement was different (R340S vs. R340H). Cybrid study revealed that the percentage of heteroplasmy was correlated with complex I function and that the novel mutation caused a much more deleterious effect than the np 11,778 LHON mutation in complex I activity. .COPYRGHT. 2003 Elsevier Science B.V. and Mitochondria Research Society. All rights reserved.

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(FILE 'HOME' ENTERED AT 11:07:07 ON 01 DEC 2006)

FILE 'MEDLINE, CAPLUS, BIOSIS, EMBASE, SCISEARCH' ENTERED AT 11:07:50 ON 01 DEC 2006

E GUY JOHN/AU  
L1 129 S E3, E4, E5, E6, E7, E8, E9  
L2 23 S L1 AND MITOCHONDRIA  
L3 7 S L2 AND ND4  
L4 8 S ND4 (A) MITOCHONDRIA  
L5 2046 S (LEBER HEREDITARY OPTIC NEUROPHY) OR LHON  
L6 391 S L5 AND MITOCHONDRIA  
L7 101 S L6 AND ND4  
L8 9 S L7 AND CODON  
L9 23 S L3 OR L4 OR L8  
E MANFREDI GIOVANNI/AU  
L10 130 S E3  
E SCHON ERIC/AU  
L11 284 S E3, E4, E5  
L12 381 S L10 OR L11  
L13 209 S L12 AND MITOCHONDRIA  
L14 6 S L13 AND ND4  
L15 13 S L13 AND CODON  
L16 19 S L14 OR L15  
L17 38 S L9 OR L16  
L18 27 DUP REM L17 (11 DUPLICATES REMOVED)